Advances in Pediatric Surgery

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• Recent advances in pediatric surgery have been made in several fields. Hydrocephalus is again being treated by draining the cerebrospinal fluid into either the ureter, the mastoid antrum or the peritoneal cavity. Funnel chest should be corrected surgically. Congenital atresia of the esophagus is best treated by a one-stage operative repair. Patent ductus should be closed. Operations are available for cyanotic children. Intussusception is again being treated by barium enema in selected cases. Megacolon can be benefited by surgical procedures, which now are directed at the distal spastic segment rather than the proximal dilated segment.

MANY OF THE ADVANCES in the field of pediatric surgery stem indirectly from increased knowledge and ability to cope with the fundamental problems of hemorrhage, shock, electrolyte losses, infection, and anesthesia. The theoretical concepts, which are common to medical as well as surgical pediatric problems, are rather difficult to implement in children, particularly in newborn infants, and certain practical advances have been made in recent years which are worthy of emphasis. These practical considerations pertain to the administration of anesthesia and parenteral solutions.

Endotracheal anesthesia in infants was occasionally followed by obstruction to the airway from edema and irritation. This may still occur, but is becoming rare now that small endotracheal catheters of inert plastic material have been developed for endotracheal intubation of infants. Similarly, the control of the flow of respiratory gases through endotracheal tubes has been facilitated by the use of non-resistance valves (Slater valves). The value of bronchoscopy and its use in aspiration of the tracheobronchial tree for the prevention and treatment of atelectasis has become available for newborn infants with the development of a bronchoscope and laryngoscope (Michelson[10]) especially for infants.

The best treatment of shock is prevention, and the simplest manner of preventing fluid, blood, and electrolyte loss during major surgical procedures in infants is to insert a small polyethylene catheter into the internal saphenous vein. This is an extremely valuable procedure which can be done very simply under local anesthesia. It may be done while the child is being prepared for operation or during anesthetization immediately before operation. A transverse incision 1 cm. in length is made in the skin just anterior to the internal malleolus. The subcutaneous fat is separated bluntly in the direction of the internal saphenous vein until the vein is identified. The vein is ligatured distally, an incision made into it, a plastic catheter or a blunt-needle cannula inserted, and passed upward into the saphenous vein for a few inches. The cannula is tied in place, and a single suture closes the skin wound. It should be remembered that as it is difficult or impossible to draw blood back freely in newborn infants, the best way to make sure the indwelling catheter is functioning is to drip fluid into the vein. The catheter may be used for continuous drip infusion in the postoperative period for several days. This simple and satisfactory method of parenteral intubation is a significant advance in pediatric surgery and may prove life-saving.

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NEUROSURGICAL ADVANCES

The surgical treatment of hydrocephalus has occupied the interest and attention of pediatricians and surgeons alike. Hydrocephalus may be related to a block in the free circulation of the cerebrospinal fluid, either on a congenital, traumatic, or postinflammatory basis; or it may result from failure of absorption of the cerebrospinal fluid. Diagnostically, dye studies and air studies to determine the type of hydrocephalus are particularly important, for upon the diagnosis depends the kind of surgical procedure to be employed.

Torkildsen's Procedure.[15] The Torkildsen procedure consists in inserting a rubber catheter or polyethylene tubing into the posterior horn of one of the lateral ventricles, passing the tubing beneath the bone but outside the dura, and implanting the other end into the posterior cistern. It is also possible to pass the shunting tube subcutaneously. This shunt relieves an obstruction to the intracranial flow of cerebrospinal fluid, and is applicable to tumors of the pineal gland, third ventricle, aqueduct, or fourth

ventricle. It also by-passes a congenital stenosis of the aqueduct of Sylvius. It requires a normal posterior cistern and a normal absorption of cerebrospinal fluid once it has gained access to the posterior cistern. It is not applicable, therefore, to the obstructive hydrocephalus often associated with spina bifida, which is generally owing to an accompanying Arnold-Chiari malformation.

Matson Procedures.[8] In 1951 Matson reported the successful use of two procedures in hydrocephalus, the choice between them depending upon the presence or absence of a block to the free flow of cerebrospinal fluid:

1. *Anastomosis between the lumbar subarachnoid space and the ureter.* If no obstruction exists to the flow of cerebrospinal fluid, as in the communicating or idiopathic hydrocephalus, a shunt can be established between the lumbar subarachnoid space and the ureter. This consists in sacrificing one kidney, using a small polyethylene tube to bridge the gap, and connecting the lumbar subarachnoid space to the ureter.

2. *Ventriculo-ureterostomy.* In patients who have an obliteration of the spinal as well as the intracranial surface subarachnoid spaces the previously described shunting procedures would be useless. Matson,[9] therefore, has elected to shunt the cerebrospinal fluid directly from the ventricle into a ureter by means of a long subcutaneously-implanted polyethylene tube. The procedure seems indicated in hydrocephalus of the following types:

(1) Obliteration of the absorptive surfaces for the cerebrospinal fluid, as an aftermath of purulent meningitis or diffuse hemorhage into the subarachnoid space. Many more patients survive purulent meningitis nowadays with antibiotic therapy, and obliteration of the posterior cisterns and subarachnoid absorptive pathways is more frequently encountered.

(2) In benign obstructions of the aqueduct of Sylvius or posterior fossa, either on a congenital basis or from Arnold-Chiari malformation. The subarachnoid pathways may also be obliterated or may not have developed. In such circumstances, ventriculo-ureterostomy offers relief from progressive hydrocephalus.

In young children the obvious problem with this procedure is that of subsequent growth. The consequences of this difficulty will have to be evaluated in the future.

It should also be emphasized that in the immediate postoperative period problems in fluid and electrolyte imbalance are likely to be troublesome. Attention must be given to chloride, sodium, water and potassium balance.

Nosik Procedure[11] (ventriculomastoidostomy). The indications for this procedure are similar to those for ventriculo-ureterostomy. It has the advantage of avoiding fluid and electrolyte loss from the body, for the fluid passes from the ventricle into the mastoid antrum, then into the middle ear and the eustachian tube, and finally is swallowed from the nasopharynx. It also creates a pathway between the ventricle and nasopharynx, a possible route for retrograde infection. In practice, however, the differential pressures and the action of the cilia in the eustachian tube seem to minimize the hazard of retrograde infection.

Briefly, the procedure consists in shunting the cerebrospinal fluid from the lateral ventricle into the mastoid antrum by a small polyethylene tube.

None of the various shunting operations for hydrocephalus is applicable or justified once irreparable brain damage has already occurred. They are not a means of prolonging a demented life.

THORACIC OPERATIONS

Advances in both techniques and concept of thoracic surgery have been made in recent years. Technical advances have permitted the surgical repair of such conditions as funnel chest, atresia and stricture of the esophagus, patent ductus arteriosus, coarctation of the aorta, ringed aorta, and anomalous subclavian vessels. Conceptual advances have resulted in the use of operations to alter the flow of blood through the lungs; and cyanotic heart disease, pulmonary stenosis, and transposition of the great vessels have become amenable to surgical benefit.

Funnel Chest. The cause of funnel chest is still not clear, but it is related to a short central tendon of the diaphragm in infancy and in later years to a progressive deformity in the growth of the sternum and attached costal cartilages. Surgical correction is advisable in order to correct the deformity, which becomes of psychological concern to a growing child, to prevent progressive increase of the deformity with attendant kyphosis and rotation of the sternum and costal structures, and to relieve cardiac and pulmonary embarrassment which may occur in later years from compression of the heart or lungs.

It is not possible to predict the rate of progression of the deformity. In general, the earlier surgical correction is initiated the simpler the surgical procedure required. In cases of long standing, with severe deformity in adolescence, extensive operation may be only partially corrective.

In young infants and in children under two years of age the most commonly used surgical procedure is to section the attachment of the central tendon of the diaphragm to the xiphisternum after excision of the xiphoid process. The subcostal ligament is

also incised and the sternum elevated. The release of the abnormal pull of the diaphragm on the sternum permits elevation of the sternum and prevents or retards subsequent progression of the deformity with the attendant costal arch deformity. In older children, or in adolescents, it is generally necessary to excise all the deformed costal cartilages and ribs anteriorly from the second to the seventh inclusive, mobilize the sternum from the anterior mediastinal structures, osteotomize the sternum at the sternomanubrial junction, and then maintain the sternum in an elevated position until healing has occurred either by suturing the sternum to the manubrium in the corrected position or by using a long rib strut beneath the sternum to maintain proper elevation.

Congenital Atresia of the Esophagus. The commonest type of congenital atresia of the esophagus, which results from a failure of proper embryonic separation of the esophagus from the tracheobronchial tree, is as follows: The upper end of the esophagus terminates as a dilated blind sac about the level of the first to second thoracic vertebrae, and the lower end of the esophagus, which is thin and undeveloped, communicates with the trachea through a tracheo-esophageal fistula. In former years it was possible to reconstruct the defects by employing staged operations and eventually obtain continuity of the upper alimentary tract by an antethoracic cervical esophagogastrostomy. This entailed months of hospitalization, often with a distressing result from the parental viewpoint, even if the child survived. The brilliant advances in thoracic surgery in infants in recent years now permit a one-stage reconstruction of this disorder by direct repair of the tracheo-esophageal fistula and a simultaneous end-to-end anastomosis of the esophageal segments. A normal anatomical and physiological alimentary tract is established. The mortality rate from the procedure is less than 50 per cent, taking statistics the country over. The sooner operation is performed, the less likelihood of attendant, oftentimes fatal, complications from pulmonary infection, dehydration, and malnutrition. It is imperative, therefore, that the condition be recognized early. Inability to swallow, respiratory distress and cyanosis on feeding, and the spilling over of food or saliva into the tracheobronchial tree are symptoms. The diagnosis may be readily confirmed by inability to pass a catheter down the esophagus and by roentgen studies made with Lipiodol® to outline the upper blind sac of the esophagus. The presence of air in the intestinal tract establishes the presence of a tracheo-esophageal fistula in association with the upper blind esophageal sac.

The author's preference is for right transthoracic extrapleural repair with resection of a segment of the fourth rib. In this procedure the lower end of the esophagus is disconnected from the trachea, the tracheal fistula repaired, and a primary end-to-end anastomosis of the esophageal segments is done. Gastrostomy is established for feeding until adequate food can be taken orally.

Stricture of the Esophagus. Congenital complete atresias of the esophagus can be repaired by antethoracic cervical esophagogastrostomy. When the patient is older it is possible to drop the antethoracic stomach back into the thoracic cavity to improve the cosmetic result. Localized congenital strictures or diaphragms of the esophagus can be repaired by transthoracic resection of the esophagus with primary end-to-end anastomosis of the esophagus. Extensive strictures can be corrected by resection of the strictured area and intrathoracic esophagogastrostomy, a procedure that entails sacrifice of the cardiac sphincter and may be followed by peptic esophagitis.

The commonest strictures of the esophagus, however, result from trauma—usually from swallowing of a caustic material such as lye. In recent years the armamentarium for management of such lesions has been enhanced by the development of the Tucker[16] indwelling dilators and the Hurst mercury bougies. Once satisfactory dilation of the stricture has been obtained, the patient or the parents can be taught the use of the Hurst mercury dilators to maintain the esophageal lumen.

Patent Ductus Arteriosus. Patent ductus arteriosus was the first congenital lesion of the great vessels to be treated successfully, and universal experience since 1938 has attested the success and safety of the operation first performed by Gross.[4] A child in whom the condition exists may appear to be normal, or he may be underdeveloped. A palpable thrill over the precordium and a continuous, harsh "machinery" murmur of an arteriovenous fistula, heard best over the second and third interspaces to the left of the sternum, are characteristic. There is greater than normal disparity between systolic and diastolic blood pressure, with the diastolic pressure low. As much as 70 per cent of the blood may be shunted through the ductus back into the lungs, and in x-ray studies an enlargement of the pulmonary conus and increased vascular markings in the lung fields may be noted.

If a ductus does not close by the age of two years operation should be advised, for the period in which operation can be most easily and safely accomplished is between two and six years of age. The surgical procedure consists in division of the ductus, if this can be performed safely; or, if the ductus is so short and broad that division seems unsafe, multiple suture ligatures may be used with only a rare recurrence.

Coarctation of the Aorta. The diagnosis of coarctation of the aorta is ordinarily not difficult, if the condition is suspected. Nosebleeds, headaches, cold legs, pains in the extremities, and easy fatigability on running should arouse suspicion of this lesion. Hypertension in the upper extremities with femoral pulsations and pressure diminished or impalpable affords a clinical diagnosis, which can be verified by roentgenographic observation of notching of the ribs and left ventricular hypertrophy. There are two common types: (1) the adult type, in which the constriction is short, and in which resection of the coarcted area with end-to-end anastomosis of the vessel is feasible; and (2) the infantile type, in which the coarcted segment is long and irregular, oftentimes with a patent ductus arteriosus entering below the coarctation, and in which direct repair of the aorta is not possible.

Crafoord[3] and Gross[5] described the successful repair of coarctation of the aorta in patients by direct end-to-end anastomosis. The pertinent remaining questions as to growth of anastomosed vessels after circumferential suturing and as to the technique of bridging aortic defects have been partially answered in recent years. It has been observed that the size of an anastomosed area increases somewhat with the size of the patient, particularly if the suture line is interrupted in several places in its circumference. Aortic defects have been bridged by direct substitution of specially preserved aortic homografts (Gross[6]), and other investigators have circumvented the problem by swinging the left subclavian vessel down to bridge the gap and carrying out anastomosis between it and the aorta.

As there is little likelihood that coarctation will cause serious damage before the age of six years, the optimum age for operation seems to be between six and fifteen years. As three-fourths of patients with coarctation of the aorta die in early adult life, operation before age fifteen is advisable.

Anomalous Subclavian Artery and Ringed Aorta. Anomalies of the arch of the aorta are not uncommon, but not all of them cause symptoms. Symptoms, if they occur, are related to respiratory or esophageal obstruction. Difficulties in swallowing or repeated bouts of dyspnea, respiratory distress, or coughing should arouse suspicion of these lesions, particularly if other congenital anomalies exist. The diagnosis is confirmed by x-ray visualization of compression of the esophagus. The two commonest anomalies of the arch are ringed aorta and aberrant right subclavian artery. In repair of ringed aorta the smaller aortic arch is divided and the ends sutured. Anomalous right subclavian artery is corrected by dividing the artery just distal to its origin from the descending aorta and permitting it to retract to the right of the trachea and esophagus.

Cyanotic Heart Disease. Congenital heart disease is commonly divided into the cyanotic and acyanotic groups. Any condition which permits shunting of blood from the right side of the heart to the left, or which prevents an adequate flow of blood through the lungs will result in incomplete saturation of the arterial blood. Cyanosis results, or rather is "visible," when the amount of oxygen-deprived hemoglobin in blood circulating in the "visible" capillary beds (lips, fingernails) is 5 gm. or more per 100 cc. In children, cyanosis from failure of blood to be oxygenated as it passes through the pulmonary bed is rare, and the common cause of cyanosis is from venous admixture. The common cyanotic lesions which have become amenable to surgical benefit are:

1. *Tetralogy of Fallot:* The four characteristics of this anomaly are pulmonary stenosis, interventricular septal defect, dextroposition of the aorta which overrides the septal defect, and hypertrophy of the right ventricle.

A patient with this condition is cyanotic, and in x-ray films of the chest diminished pulmonary artery markings and right ventricular enlargement may be observed. The general principle of surgical treatment is to increase the amount of blood flowing through the pulmonary circuit. In infants, creation of this life-saving artificial ductus is best accomplished by direct anastomosis of the aorta to the pulmonary artery, the Poth-Smith operation.[12] In children over two years of age the Blalock-Taussig operation[1] is preferable, namely, anastomosis of the subclavian artery to the pulmonary artery. The optimal age for the operation seems to be between the ages of five and ten, but it may have to be done earlier if life and development are unduly threatened at an earlier age. "The sicker the sooner" is the accepted motto.

Some day it may be possible to deal with the problem more directly by surgical enlargement of the stenosed portion of the pulmonary artery, but this must await further refinements in angiocardiography to permit better visualization of the existing anomalies and a greater experience with the technique of Brock[2] for direct valvulotomy. It is probable, however, that the direct method would not be applicable if the stenosed portion was long, but if it was short and diaphragm-like and the pulmonary artery beyond it was enlarged, direct operation probably would increase the flow of pulmonary blood without adding another cardiovascular anomaly.

2. *Stenosis of the pulmonary valve.* This condition is statistically one-tenth as common as the tetralogy of Fallot. It is accompanied by cyanosis in 70 per cent of cases because of accompanying interauricular septal defect or interventricular septal defect with a right-to-left shunt. Dilation of the pulmo-

nary artery beyond the point of stenosis, which is characteristic, may be observed on a plain film of the chest or angiocardiographically. Right ventricular pressure is very high, and the right ventricular hypertrophy observed at operation is striking. If the pulmonary artery is palpated at a point immediately beyond the site of stenosis, the spurting of the jet-like stream of blood through the stricture can be felt. Creation of an artificial ductus in this situation may initiate right-sided heart failure. The operation of choice is the Brock procedure,[2] in which the pulmonary valve is cut and dilated through the use of special valvulotomes and dilators passed through a small incision in the right ventricle.

Other Less Common Anomalies. Surgical possibilities are being explored in the management of complete transposition of the aorta and pulmonary vessels and in the treatment of tricuspid atresia. Results in these conditions are still quite unsatisfactory. Similarly, the closure of interauricular and interventricular septal defects awaits the development of suitable pump-oxygenators which will sustain both cardiac and pulmonary functions while intracardiac operation is being carried out.

ABDOMINAL OPERATIONS

Intussusception. In recent years there has been a renewal of interest in nonoperative management of intussusception. The majority of cases of intussusception occur in infants under two years of age, and rarely is a local lesion such as a polyp, tumor, Meckel's diverticulum, or ectopic pancreatic nodule encountered. Intussusception generally begins at the ileocecal valve, and as the intussuscepted bowel drags more of the enveloping loop in after it, the head of the intussuscipiens remains constant. The most characteristic symptom is discharge of blood into the bowel owing to the vascular embarrassment of the intussusceptum. Edema and compression narrow the lumen of the bowel, causing first partial and later complete obstruction. Gangrene of the bowel occurs after a variable period of time, depending upon the degree and completeness of vascular occlusion. Oftentimes a sausage-shaped mass is palpable in the left upper quadrant of the abdomen and there is absence of bowel in the right lower quadrant. Colic and vomiting, blood in the stool or discharge of bloody material, partial obstruction, and an abdominal mass occur early; later, ileus, peritoneal irritation, and manifestations of toxicity appear. The diagnosis is confirmed by roentgen examination after barium enema.

In clinical and experimental observation[13] it has been noted that barium introduced under hydrostatic pressure of three to three and one-half feet will usually reduce non-gangrenous intussusception but will not reduce gangrenous intussusception nor risk rupture of the intussuscipiens or intussusceptum. Accordingly, conservative management consists in inserting a Foley catheter into the rectum, holding the buttocks together, and running in barium under hydrostatic pressure of three feet. If complete reduction of the intussusception occurs, the intussuscepted loop will disappear and barium will break through the ileocecal valve into the terminal ileum. No further treatment is then necessary, and recurrence of the intussusception is extremely rare.

If the intussusception cannot be reduced by hydrostatic pressure with barium, or if any doubt exists as to the completeness of the reduction, or if there are continuing symptoms of peritoneal irritation to cause suspicion of the viability of the intussuscepted bowel, immediate laparotomy is indicated. This can ordinarily be accomplished through a McBurney or short transverse right-sided incision.

Hirschsprung's Disease (megacolon) : Concepts of the cause and treatment of megacolon are in process of revision, and the condition is no longer regarded as an idiopathic dilation of the colon. Formerly, cases in which intestinal dilation and severe constipation were present and in which a tremendously dilated colon with poor haustral markings was observed in roentgen studies with barium enema were grouped as "idiopathic megacolon." It now seems better to divide such cases etiologically:

1. *Achalasic or aganglionic megacolon* (true Hirschsprung's disease). In this condition the parasympathetic ganglion cells of the myenteric plexus are absent. The aganglionic area, usually the lower rectum and the sigmoid colon, is narrow and spastic. The colon is greatly dilated above the aganglionic segment. Usually the anus and terminal rectum are normal.

2. *Functional megacolon.* In megacolon of this classification, the colonic dilatation is secondary to functional obstruction in the bowel. The origin of this functional obstruction may be dietary, psychologic (improper bowel training), endocrinologic (hypothyroidism and cretinism), or pain and spasm from an anal fissure.

3. *Organic megacolon* is that which is caused by an anal stricture, by inadequately repaired anal atresia, or by extrinsic or intrinsic bowel tumor that mechanically obliterates the lumen of the bowel.

In functional and organic megacolon the cause is apparent and the treatment is to eliminate the cause.

As to aganglionic megacolon, data obtained through pathological examination of the diseased segment and results of studies of physiological pressure with intracolonic balloons have led to a changed concept of therapy. Whereas previously the dilated

segment was looked upon as the diseased segment, and was resected (oftentimes after a preliminary colostomy proximally), nowadays attention is directed to resecting the aganglionic distal narrow segment as a means of relieving the megacolon. Svenson[14] and Hiatt,[7] both advocates of this concept, have developed a sphincter-saving operation which permits resection of the diseased aganglionic segment with preservation of the anus and its sphincter. In that procedure the ganglion-containing dilated portion of the bowel above the resected narrowed segment is brought down through the anal sphincter and sutured to the anal mucosa. Further time will be necessary to evaluate late results, but the immediate results have been highly satisfactory.

It is worthy of emphasis that this operation is applicable only if the megacolon is of aganglionic type. Diagnosis of this condition is best established by noting the onset of obstructive symptoms, which is invariably at birth or within the first few days thereafter. The narrow obstructed rectosigmoid is best visualized roentgenographically in oblique films taken after the large redundant sigmoid colon has been manipulated out of the pelvis with the patient in the Trendelenburg position. Svenson emphasized that, since many patients with aganglionic megacolon also have an atonic bladder, cystometrograms with residual urine determinations are advisable preoperatively.

Once the diagnosis of true congenital megacolon has been made, operation should be considered if the roentgenographic characteristics are present or if the disease is progressive. For extremely sick patients, preliminary colostomy may be necessary; otherwise, a one-stage procedure is safe and satisfactory.

How some patients with megacolon were cured previously by resection of the dilated segment is subject for speculation.

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